

Clinical Article

Epidermoid Tumors in the Cerebellopontine Angle Presenting with Trigeminal Neuralgia

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Objective : The purpose of this study is to evaluate the clinical characteristics and surgical outcome of cerebellopontine angle (CPA) epidermoids presenting with trigeminal neuralgia.

Methods : Between 1996 and 2004, 10 patients with typical symptoms of trigeminal neuralgia were found to have cerebellopontine angle epidermoids and treated surgically at our hospital. We retrospectively analyzed the clinico-radiological records of the patients.

Results : Total resection was done in 6 patients (60%). Surgical removal of tumor and microvascular decompression of the trigeminal nerve were performed simultaneously in one case. One patient died due to postoperative aseptic meningitis. The others showed total relief from pain. During follow-up, no patients experienced recurrence of their trigeminal neuralgia (TN).

Conclusion : The clinical features of TN from CPA epidermoids are characterized by symptom onset at a younger age compared to TN from vascular causes. In addition to removal of the tumor, the possibility of vascular compression at the root entry zone of the trigeminal nerve should be kept in mind. If it exists, a microvascular decompression (MVD) should be performed. Recurrence of tumor is rare in both total and subtotal removal cases, but long-term follow-up is required.

KEY WORDS : Epidermoid · Cerebellopontine angle · Trigeminal neuralgia · Microvascular decompression.

INTRODUCTION

Intracranial epidermoids are rare, histologically benign, slow-growing, congenital neoplasms of the central nervous system that may arise from retained ectodermal implants^{1,11,16,17,21,23}. Epidermoid tumors are the most common of all embryonal intracranial neoplasm and account for approximately 1% of all intracranial tumors^{1,11,16,17,21,23}. The cerebellopontine angle (CPA) is one of the most common sites affected^{19,23}. Epidermoids can cause irritation of the cranial nerve resulting in cranial nerve hyperactive dysfunction, such as trigeminal or glossopharyngeal neuralgia, or hemifacial spasm^{11,12,16} and also may present symptoms of cranial nerve, cerebellar and brain stem dysfunction, as

well as hydrocephalus and meningeal irritation^{16,19,21,23}. The incidence of symptom and the anatomic relationship between tumor and the respective cranial nerve have been described in previous reports^{4,5,11}. To those patients who present with complaints of trigeminal neuralgia, the long-term prognosis for post-operative pain relief is of particular interest, but little is known about long-term surgical results.

We retrospectively reviewed and analyzed the clinical characteristics of patients with trigeminal neuralgia due to CPA epidermoid. We also compared their characteristics with trigeminal neuralgia due to vascular compression. Our review was focused on the clinical characteristics, operative findings and long-term surgical results of CPA epidermoids presenting with trigeminal neuralgia (TN).

MATERIALS AND METHODS

This study represents a retrospective review of patients treated for cerebellopontine angle epidermoid tumors with trigeminal neuralgia between 1996 and 2004. For inclusion in this study, all patients had to fulfill the diagnostic criteria of

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Neuroradiological investigation

All of the patients were preoperatively evaluated with magnetic resonance imaging, including 3 Dimensional Fourier Transformation-Constructive Interference in Steady State (3 DFT-CISS) sequences, which provide images with high spatial resolution and good contrast between solid structures and the CSF. On the MR studies, epidermoids mostly displayed hypointensity on T1-weighted images, hyperintensity on T2-weighted images, and no enhancements after contrast material injection. The CISS sequences depicted all tumors as hypointense relative to cerebrospinal fluid. Because 3-D CISS sequences provided high spatial resolution with very thin section, it was possible to detect the exact tumor extension, even for a small lesion in Meckel's cave (Fig. 1). With the use of MR imaging, correct diagnosis of epidermoid tumors could be made preoperatively in all cases.

Operation

A lateral suboccipital approach through retromastoid craniectomy was performed in 9 patients (one bilateral CPA epidermoid cyst case was treated by staged bilateral retromastoid craniectomy). For the remaining one patient, who had a supratentorial extension and preoperative hearing loss, a retrolabyrinthine presigmoid approach was used (Table 3).

After opening the dura, the arachnoid was divided and the tumor capsule was exposed. Its remarkable white and pearly appearance made immediate confirmation of an epidermoid. In 6 cases, the trigeminal nerves were completely encased by tumor without displacement. The nerve was compressed and distorted by the epidermoid in 3 patients. In 1 patient, the anterior inferior cerebellar artery (AICA), located on the same side of the tumor in relation to the trigeminal nerve, was found to be compressing the nerve directly on its ventrocaudal aspect; thus, the nerve was compressed by both the tumor and the artery on the same side. In this patient, MVD of the trigeminal nerve was performed in addition to tumor removal (Fig. 2). The usual techniques of intracapsular tumor removal and extra-capsular dissection were applied to remove the epidermoid tumor. An important concept to bear in mind during debulking was that the nerves and vessels were embedded in the tumor and could be densely adherent to it. During the operation, irrigation and cotton pads were used to reduce spillage of the irritating cyst content into the subarachnoidal space. Total resection of the epidermoid content and radical resection of the cyst capsule were performed in 6 patient

Table 2. Comparison of clinical features in patients with trigeminal neuralgia resulting from an epidermoid and vascular cause

Patient data	Epidermoid cases	Vascular compression cases	Significance
No. of affected division	1.4 ± 0.516	1.51 ± 0.504	$p = 0.5258$
Trigger point (presence : absence)	8 : 2	52 : 9	$p = 0.305$
Age at operation (yr)	46.8 ± 12.0904	57.90 ± 11.262	$p = 0.00056$
Age at onset (yr)	42.95 ± 10.89	52.51 ± 11.770	$p = 0.0189$
Duration of symptoms (yr)	3.72 ± 4.521	5.39 ± 4.245	$p = 0.2569$

Table 3. Surgical approach and extent of removal in cerebellopontine angle epidermoids

Surgical method	No. of cases
Surgical approach	
Retromastoid	9
Retrolabyrinthine presigmoid	1
Extent of removal	
Total	6
Subtotal & left capsule	4

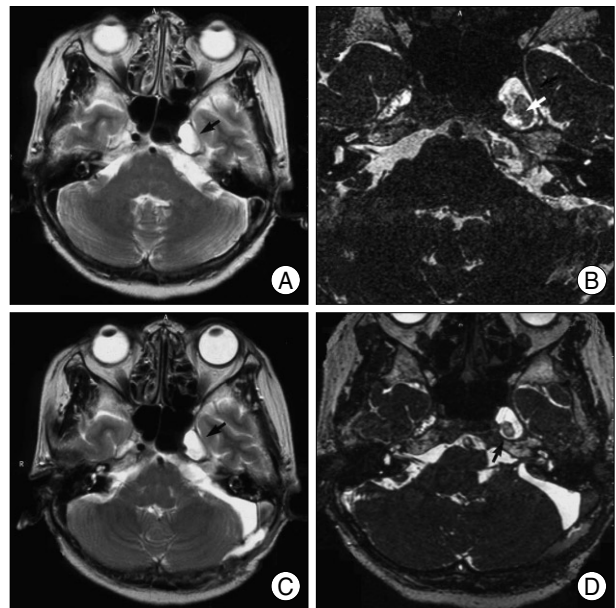


Fig. 1. Large left CP angle epidermoid cyst displays hyperintense signal in T2 weighted image. The lesion extends into the Meckel's cave (arrow) and compresses the ipsilateral pons (A). Preoperative CISS image distinguish the lesion from CSF space (B). Postoperative axial T2 weighted image (C) and CISS image (D) show the remnant mass in the Meckel's cave (arrow). CISS : Constructive Interference in Steady State, CP : cerebellopontine, CSF : cerebrospinal fluid.

(60%). In 4 patients (40%), the cyst capsule or a small amount of tumor content were left because of dense adhesion to the critical neurovascular structure or tumor extension beyond the midline (Table 3).

Surgical results and complications

All patients showed immediate relief of TN after surgery. Symptoms of facial paresis and hearing disturbance the

Table 4. Operative outcome and follow-up results in epidermoids

Operative method	Case	Outcome			Follow-up results	
		Good	Deficit	Death	Mean follow-up duration(yr)	Recurrence
Retromastoid approach	9					
Total removal	6	6	0	0	5.83	No
Subtotal & left capsule	3	3	0	0	9	No
RLPS approach	1					
Total removal	0	0	0	0	-	-
Subtotal removal	1	0	0	1	-	-

RLPS, retrolabyrinthine presigmoid

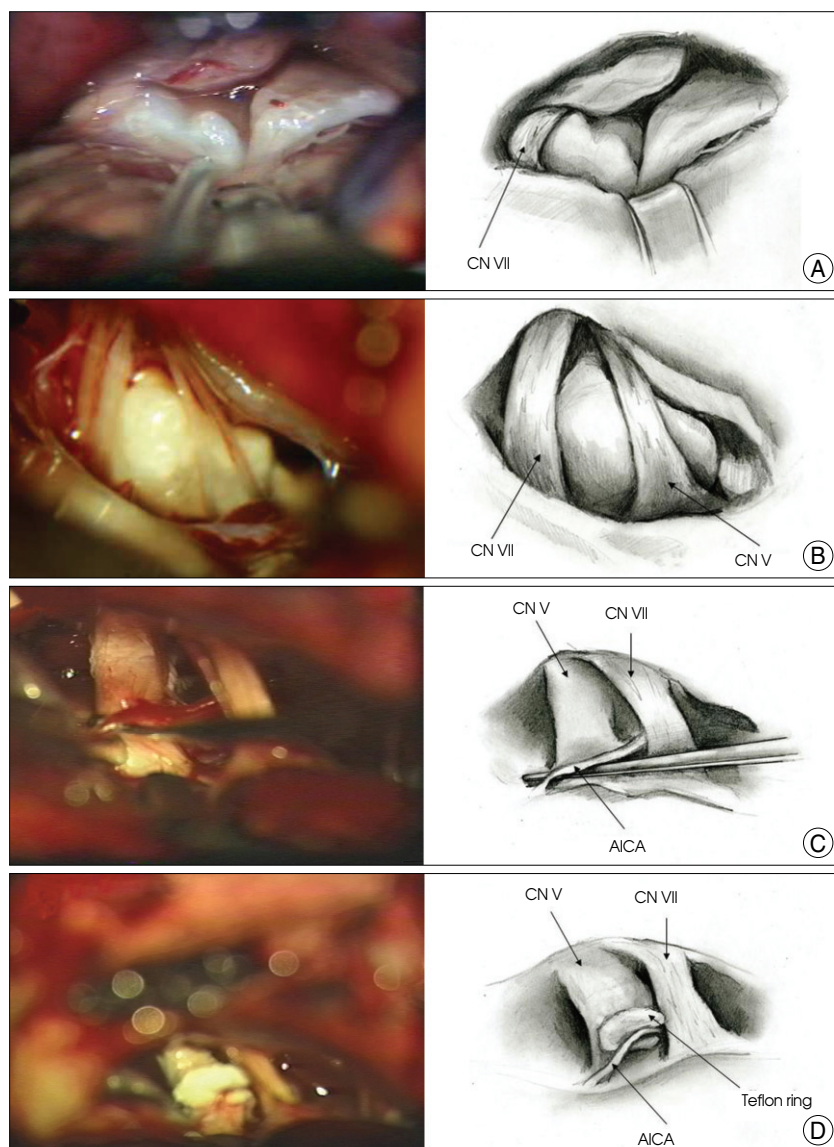


Fig. 2. Photographs and schematic drawings displaying the anatomic relationship among the CPA epidermoid, trigeminal nerve, and the artery. A : The trigeminal nerve is completely encased by the epidermoid without displacement. B : During the operation, the trigeminal nerve is found to be compressed and distorted by the tumor. C and D : After tumor removal, the anterior inferior cerebellar artery (AICA), located on the same side of the tumor in relation to the trigeminal nerve, is found to be compressing the nerve directly on its ventrocaudal aspect, thus, the nerve is compressed by both the tumor and the artery on same sides (C). In this patient, microvascular decompression (MVD) of the trigeminal nerve in addition to tumor removal was performed (D).

patients experienced prior to the surgery did not show any evidence of improvement, but diplopia (CN VI deficit) gradually improved.

Surgical outcomes were divided into the 3 groups of 'good' (for cases with no cranial nerve deficit), 'deficit' (for cases with cranial nerve deficit) and 'death' (for cases of patient's death). Nine of the ten patients recovered with no post-operative complications, and one deceased due to chemical meningitis, cerebellar and brain stem infarction (Table 4). All patients were followed up with magnetic resonance (MR) imaging and computed tomography (CT) for early detection of tumor recurrence. In particular, patients with a residual tumor were followed up with MR imaging every 6 months for 2 years, and yearly thereafter. During mean follow up of 6.8 years after surgery, no peculiar symptoms set in, nor did the trigeminal neuralgia recur. As for the one case in which tumor content was removed subtotally and two cases in which capsules were left, remnant tumors were detected by postoperative MR, but there was no regrowth of tumor. Six cases of complete removal also showed no evidence of recurrence.

DISCUSSION

Epidermoid tumors represent 0.2 to 1.4% of all primary intracranial tumors^{11,15,16,21}. CPA epidermoids constitute 40% of all intracranial epidermoids¹⁶ and the incidence of epidermoids among all CPA tumors is approximately 5%¹¹. Incidences of TN in patients with a CPA epidermoid has been reported to vary from 0 to 90.6%^{4,5,11,20,24}. But the incidence of CPA epidermoids in patients with TN has also been variously reported as 0.2 to 5.5% in the literature^{4,11,18}. In our series, all patients with a CPA epidermoid were associated with TN, per-

haps because all of these patients were referred to our department for treatment of facial pain.

Epidermoid tumors have an extremely slow linear growth rate². Therefore, symptoms are long-lasting and patients may display symptoms late in the course of the illness. However, patients often present relatively early when the predominant symptom is trigeminal neuralgia¹⁶. The symptoms and signs are caused by displacement of the adjacent neurovascular structure. It has been reported that hearing loss is the most common symptom in patients with epidermoids, accounting for 37.6%, followed by TN, 29.7%; dizziness or vertigo, 19.4%; facial palsy, 19.4%; headache, 17.9%; and diplopia, 16.7%¹¹. Baker et al.⁴ and Jannetta⁹ reported that patients with a tumor as the cause of TN have a clinical history that is not different from patients with classic manifestations of TN. TN due to CPA epidermoids may be clinically indistinguishable from TN with a vascular cause^{11,15}. But the occurrence of TN at a younger age and the long duration of symptoms are characteristic of TN patients with epidermoids, in contrast with TN due to vascular causes¹¹. This can be explained by the extremely slow growth rate of the tumor². The results of our study also suggest that the early onset of TN is characteristic of TN due to an epidermoid, but there was no significant difference in the preoperative duration of symptoms. In young patients with TN, therefore, the neuroimaging study must be performed carefully to avoid overlooking a small tumor in the CPA cistern.

The pathogenetic mechanism of TN in patients with CPA epidermoid remains uncertain. Direct compression of the nerve at the root entry zone^{6,17}, displacement of the trigeminal nerve and compression against a blood vessel^{5,9} at the root entry zone, or a combination of the two³ have been postulated to be the cause. The irritant, inflammatory nature of the epidermoid tumor also has been suggested to cause pain where the nerve is completely wrapped by the tumor¹⁹. The keratin contents may directly cause irritation and hyperactivity of the nerve without an intervening vascular loop²¹.

Magnetic resonance imaging is the diagnostic modality of choice for detection of an epidermoid. Nevertheless, differentiation between arachnoid and epidermoid cysts is still sometimes difficult. On standard T1- and T2-weighted spin-echo MR images, epidermoids can often barely be distinguished from CSF^{8,10}; therefore, other imaging sequences have been investigated. Fluid-attenuated inversion recovery (FLAIR) imaging, diffusion-weighted imaging, and CISS sequencing have been recommended for the characterization of epidermoid cysts^{8,10,14}. In our experience, a CISS image is the best sequence for detecting epidermoids. Although most epidermoid tumors are slightly hyperintense relative to CSF on

T1-weighted image¹⁰, CISS sequences demonstrate these lesions more clearly⁸. Because a CISS sequence provides high spatial resolution with very thin section, it is possible to detect the exact tumor extension, even for a small lesion in Meckel's cave⁸.

The optimal surgical treatment is radical removal of the tumor capsule and, for the reason that the capsule in place appears to guarantee recurrence^{1,24}. The rates of complete removal in patients with CPA epidermoid have been reported to be 18% to 97%^{16,19,21,23,24}. However, the peculiar growth feature along with the wide extension of the tumor and its firm adhesion to critical neurovascular structures make total surgical removal not always possible nor advisable^{5,16,21}. We agree that the ideal goal in surgery for epidermoids is total removal, but not at the expense of neurological deficits. It is our strategy to attempt a total removal of the capsule. If the capsule is firmly adherent to critical neurovascular structures, we leave the adherent portion in place to minimize the risk of neurological sequelae. Although a capsule remnant probably will result in recurrence, this will occur many years after surgery. Berger et al.⁵ reported that it may take 30 to 40 years for recurrent symptoms to develop, a finding that supports the rationale for avoiding radical excision.

Tumor removal itself may relieve symptoms, but it must be emphasized that epidermoid tumors commonly cause persistent distortion of trigeminal nerve even after satisfactory resection¹. Therefore, severing and removing the adherent tumor capsule and the arachnoid membrane to achieve neural axis straightening is mandatory for the cure and preventing of recurring pain¹¹. The surgeon also must pay attention to possible arterial compression at the REZ, and such an artery should be treated by MVD in addition to tumor excision¹¹. Kobata et al.¹¹ classified the relationship between the CPA epidermoid and neurovascular structure into four types according to the compression pattern of the REZ of the cranial nerve. In type A, the nerve is completely encased by the tumor without displacement of the nerve axis. In type B, the nerve is compressed and distorted by the tumor. In type C, the nerve is displaced and compressed by the artery on the opposite side of the tumor, resulting in its being pinched by both the tumor and the artery. In type D, the nerve is compressed by both the artery and the tumor from the same direction with direct contact with the artery. They advocated that MVD should be performed along with tumor removal in type C and type D. In our series, 6 (60%) case were type A, 3 (30%) were type B and 1 was type D. In type D patient, MVD was achieved in addition to tumor removal.

A relatively high incidence of postoperative cranial nerve

dysfunction has been mentioned in the previously reported series, but postoperative cranial nerve palsies tended to improve in most cases during the follow-up period^{5,20,22,24}. In our series, there was no increased postoperative cranial nerve dysfunction because we left the tumor capsule behind when the capsule was densely adherent. An operative complication unique to this type of lesion is aseptic meningitis, which is related to cholesterol crystals being spilled into the cerebrospinal pathway^{16,21-23}. Only one case occurred in our patients and the patient deceased due to chemical meningitis, cerebellar and brain stem infarction. Excision of the capsule by sharp dissection, irrigation of the CPA cistern with hydrocortisone solution during the surgery, and delayed withdrawal of steroids in the postoperative period have been advocated as possible measures for preventing chemical meningitis^{5,12,16,21}. The same procedure was adopted in all patients in the present study. Communicating hydrocephalus may develop after one or more intense periods of meningitis, or following leakage of cyst contents from the operative field^{5,16,20,23}. In our series, there was no hydrocephalus that required CSF diversion procedures. We believe that routine perioperative administration of corticosteroids and irrigation of the operative field with normal saline and hydrocortisone solution decreases the risk of hydrocephalus developing postoperatively.

Obviously, total resection eliminates the chance of recurrence in the epidermoid tumor. However, in the literature, the overall estimated recurrence rate of epidermoids in long-term follow-up has been reported to vary from 0 to 30%, with a trend toward a higher rate in cases with a longer follow-up period^{5,19-21,23,24}. As mentioned, patients with only a small amount of residual tumor after surgical resection are at low risk for recurrence during life span. However, although these tumors are histopathologically benign, rare cases have shown a tendency toward malignant degeneration, particularly to squamous cell carcinoma^{13,20}. Overall, with a median follow-up of 6.8 years, there were no recurrent cases in our series. Patients with a remnant tumor can be monitored safely for asymptomatic recurrence through a noninvasive neuroimaging technique. Early detection through routine MR imaging may improve surgical outcome in recurrent cases, since the tumor are more amenable to surgery at this stage^{20,24}. However, subsequent surgery is often indicated only when the recurrent lesion is symptomatic²⁰.

CONCLUSION

TN can be a typical symptom in epidermoid cyst in the cerebellopontine angle. This tumor can be removed easily via unilateral suboccipital craniectomy and symptoms in

our patients were relieved well and there was no recurrence symptom on the follow-up period. Epidermoid patients were clinically indistinguishable from patients with trigeminal neuralgia from vascular cause, except symptom onset at a younger age. At operation, the root entry zone of trigeminal nerve should be examined for evidence of additional vascular compression. If vascular compression exists, MVD should be performed. Recurrence of tumor is rare in both total and subtotal removal cases, but long-term follow-up is required.

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